

UC Davis

Dermatology Online Journal

Title

An intensely pruritic pebbled presentation

Permalink

<https://escholarship.org/uc/item/6tp6f9c4>

Journal

Dermatology Online Journal, 24(2)

Authors

Soriano, Livia Francine
McGrath, Conn
Hawthorne, Mark
et al.

Publication Date

2018

DOI

10.5070/D3242038171

Copyright Information

Copyright 2018 by the author(s). This work is made available under the terms of a Creative Commons Attribution-NonCommercial-NoDerivatives License, available at <https://creativecommons.org/licenses/by-nc-nd/4.0/>

Peer reviewed

An intensely pruritic pebbled presentation

Livia Francine Soriano¹, Conn McGrath², Mark Hawthorne³, Arucha Linda Ekeowa-Anderson¹

Affiliations: ¹Department of Dermatology, Barts Health NHS Trust, London, U.K., ²General Medicine, Barts Health NHS Trust, London, U.K., ³Department of Histopathology, Barts Health NHS Trust, London, U.K.,

Corresponding Author: Livia Francine Soriano, Department of Dermatology, Barts Health NHS Trust, Whipps Cross University Hospital, Whipps Cross Road, London, U.K., Email: liviasoriano@doctors.org.uk

Abstract

Lichen amyloidosis (LA) is a form of primary localized cutaneous amyloidosis (PLCA) characterized by bilateral intensely itchy domed scaly hyperkeratotic papules. Lichen amyloidosis is rare and affects men more than women. It is uncommonly seen in the western world but more prevalent in Asia. These papules most typically affect the shins and occasionally the arms and torso. Lichen amyloidosis has been reported in association with autoimmune disorders and after prolonged exfoliation and friction of affected skin. We present a 40-year-old woman with LA. In LA, the characteristic histological finding is apple-green birefringence of Congo red-stained preparations observed under polarized light. However, this is not always strongly positive, as in our patient. Other findings may include eosinophilia, periodic acid-Schiff positivity, staining with thioflavin T, and metachromasia after staining with crystal violet or methyl violet. Treatment of LA is difficult and complete clinical remission is seldom achieved. Recent trials revealed beneficial outcomes with topical calcipotriol, phototherapy, acitretin, cyclophosphamide, and laser treatments. A combination of acitretin, antihistamines, topical steroids, and hydrocolloid dressings have been beneficial in our patient with LA.

Keywords: amyloidosis, lichen amyloidosis, primary localized cutaneous amyloidosis

Introduction

Lichen amyloidosis (LA) is a form of primary localized cutaneous amyloidosis (PLCA) that affects men more

than women. The age of onset is predominantly between 40-50 years. It is uncommon in the UK, but more frequent in Asian populations [1, 2]. LA is characterized by bilateral intensely itchy domed scaly hyperkeratotic papules, which can coalesce to form plaques with a 'rippled' appearance. These most typically affect the shins, and less commonly, the arms and torso. In contrast to the nodular form of PLCA, LA does not have systemic amyloid involvement. Differential diagnoses include hypertrophic lichen planus and lichen simplex chronicus (LSC), [1].

Case Synopsis

A 40-year-old woman was referred owing to a 7-year history of an increasingly itchy eruption on her shins. The severe pruritus interfered with sleep and daily activities. Otherwise she had no systemic symptoms



Figure 1. Patient photographs: (a) hyperpigmented papules with overlying scale on lower legs, (b) left lower leg up close, and (c) right elbow.

and was usually fit and well, taking no regular or over-the-counter medications. She noted that her father had a similar condition, although he had no specific diagnosis.

On examination, she had a symmetrical eruption of hyperpigmented papules on both shins (**Figure 1a, b**) resembling sheets of pebbles. Some of these papules had overlying scale. She also had fewer skin-colored papules on both elbows (**Figure 1c**). Some papules coalesced, leading to a 'rippled' appearance. Her mucosal surfaces were unaffected.

Histology examination showed epidermal acanthosis, compact hyperkeratosis, and hypergranulosis (**Figure 2a**). The dermal papillae were widened containing homogenous dull-pink deposits associated with numerous melanophages (**Figure**

2b). These were positive for cresyl violet and weakly positive but not birefringent with Congo Red (**Figure 2c, d**). Immunoperoxidase stain for CK5/6 highlighted deposits positive in the papillary dermis.

Based on the clinical and histopathological findings, a diagnosis of lichen amyloidosis was made.

Our patient was treated with a combination of acitretin 20mg once daily, mometasone ointment daily, zinc oxide paste with ichthammol bandage, and hydroxyzine with moderate improvement in her symptoms after 3 months.

Case Discussion

The pathogenesis of LA is not fully known. A popular theory is the 'keratinocyte-theory,' whereby

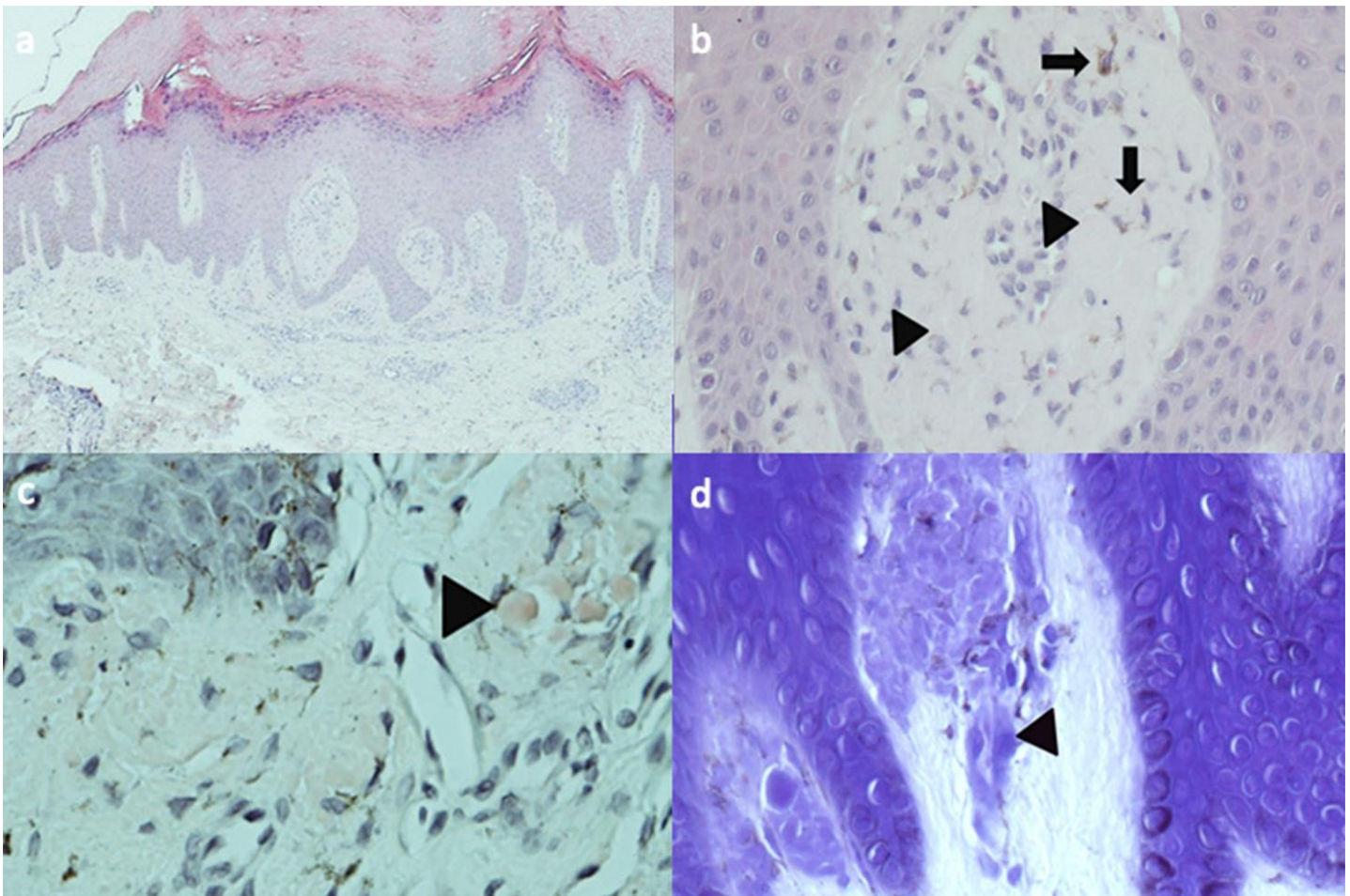


Figure 2. Histopathological findings: **(a)** Skin with epidermal acanthosis, compact hyperkeratosis, and hypergranulosis. Some of the dermal papillae appear expanded by homogenous dull-pink material. 40x; **(b)** dermal papillae containing homogenous dull-pink deposits (arrowheads) associated with numerous melanophages (arrows), H&E, 100x; **(c)** the deposits (arrowheads) were weakly positive but not birefringent on Congo red staining (400x); and **(d)** the deposits (arrowheads) were positive on cresyl violet staining (400x).

repeated trauma causes focal epidermal damage, keratinocyte degeneration, and keratin protein release. These proteins are phagocytosed by macrophages, forming amyloid deposits in the papillary dermis [3]. Prolonged exfoliation and friction from rough nylon cloth, horsehair gloves, and rough sponges have been reported to precipitate PLCA [4]. LA has been reported in association with autoimmune disorders, such as systemic lupus erythematosus, rheumatoid arthritis, Hashimoto thyroiditis, sarcoidosis, and immunoglobulin A nephropathy. Perhaps a common immune-mediated mechanism might be present in this group [5].

Confirmatory histology is required for the definitive diagnosis, although the amyloid deposits can be easily missed. In LA, amyloid is found in the papillary dermis directly below the basal epidermal layer. The overlying epidermis often shows acanthosis and hyperkeratosis, resembling LSC. There is increased pigmentation of the basal layer with corresponding melanophages in the papillary dermis. The characteristic finding is apple-green birefringence of Congo red-stained preparations observed under polarized light, a phenomenon called dichroism. However, this is not always strongly positive, such as in our case. Other findings may include eosinophilia, periodic acid-Schiff (PAS) positivity, staining with thioflavin T, and metachromasia after staining with crystal violet or methyl violet. Blood vessels remain

unaffected. Immunohistochemistry classifies the amyloid precipitates. Antibodies to epidermal cytokeratins are often found in LA. However, if staining is positive for kappa and lambda light chains, systemic amyloidosis would be suggested [1, 2].

Treatment of LA is difficult and complete clinical remission is seldom achieved. Minor trials reported in the literature revealed beneficial outcomes with topical calcipotriol, phototherapy, oral cyclophosphamide, and laser treatments [6]. Case reports have shown effective treatment with oral retinoids such as acitretin [7]. Prevention of behavior causing trauma should be addressed. A combination of acitretin, antihistamines, topical steroids, and zinc-based dressings have been beneficial in our patient with LA.

Conclusion

We share this case to remind clinicians of the striking features of lichen amyloidosis to facilitate an immediate diagnosis and appropriate management for presenting patients. Physicians must compare side effects, psychological comorbidity, and size and location of skin lesions to determine appropriate therapy for patients with lichen amyloidosis.

References

1. Schreml S. Cutaneous Amyloidoses. In: Griffiths C, Barker J, Bleiker T, Chalmers R, et al editors. *Rook's Textbook of Dermatology*. 9th ed: John Wiley & Sons Limited; 2016.
2. Kaltoft B, Schmidt G, Lauritzen AF, Gimsing P. Primary localised cutaneous amyloidosis - a systematic review. *Dan Med J*. 2013;60(11):A4727. [PMID: 24192243].
3. Schreml S, Szeimies RM, Vogt T, Landthaler M, Schroeder J, Babilas P. Cutaneous amyloidoses and systemic amyloidoses with cutaneous involvement. *Eur J Dermatol*. 2010;20(2):152-60. [PMID: 20071301].
4. Yoshida A, Takahashi K, Tagami H, Akasaka T. Lichen amyloidosis induced on the upper back by long-term friction with a nylon towel. *J Dermatol*. 2009;36(1):56-9. [PMID: 19207438].
5. Dahdah MJ, Kurban M, Kibbi AG, Ghosn S. Primary localized cutaneous amyloidosis: a sign of immune dysregulation? *Int J Dermatol*. 2009;48(4):419-21. [PMID: 19335432].
6. Weidner T, Illing T, Elsner P. Primary Localised Cutaneous Amyloidosis: A Systematic Treatment Review. *Am J Clin Dermatol*. 2017;18(5):629-642. [PMID: 28342017].
7. Vasani RJ. Response to oral acitretin in lichen amyloidosis. *Indian Dermatol Online J*. 2014;5(Suppl 2):S92-4. [PMID: 25593815].